

Balloon dilatation of the aortic valve in the fetus: a report of two cases

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Abstract

Because they had irreversible damage to the left ventricular myocardium none of 12 patients with critical aortic stenosis diagnosed prenatally survived after postnatal treatment. This experience prompted three attempts at intrauterine balloon dilatation of the aortic valve in two fetuses with this condition. On each attempt the balloon catheter was successfully delivered to the left ventricle. In the first fetus the aortic valve was not crossed and the fetus died the next day. In the second fetus the balloon was correctly positioned across the aortic valve and inflated in the valve ring. After delivery, a further balloon angioplasty was performed; this relieved the stenosis but the patient died five weeks later from persisting left ventricular dysfunction related to endocardial fibroelastosis.

Balloon angioplasty is feasible in fetal life but the prognosis depends on the ability of the relief of stenosis to limit, prevent, or allow regression of left ventricular damage before delivery.

The mildest forms of congenital aortic stenosis may become haemodynamically important only in late adult life when calcification becomes a prominent feature whereas the most severe forms are life threatening in infancy. In the neonate mortality is high and left ventricular dysfunction with endocardial fibroelastosis is common. Even when the obstruction to the left ventricular outflow tract is relieved the left ventricle often fails to maintain cardiac output.

Recently, it has become possible to detect both aortic stenosis and the severer forms of endocardial fibroelastosis in fetal life. In our unit, 28 fetuses were identified as having aortic stenosis alone (two cases) or aortic stenosis with endocardial fibroelastosis (26 cases). In two of the 12 mothers who elected to continue the pregnancy there was an intrauterine death. None of the 10 livebirths survived; only four survived long enough for treatment by balloon dilatation of the aortic valve. In addition, we saw another prenatal feature that influenced the outcome. In four of the 28 fetuses, the left ventricle failed to grow normally as gestation advanced and the left ventricle became hypoplastic which made the neonate unsuitable for relief of aortic valve obstruction.¹

Balloon dilatation is a well established technique for the relief of pulmonary or aortic

stenosis in children.^{2,3} Its efficacy in the newborn is being compared with that of surgical valvotomy.⁴ We attempted balloon dilatation in the fetus in an attempt to improve the dismal prognosis for this condition by relieving obstruction to the aortic valve before irreversible left ventricular damage had developed and to try to prevent the growth failure that we had seen. We report our experience in two fetuses in whom balloon dilatation of the aortic valve was attempted.

Patients and methods

Both patients were referred to the Department of Perinatal Cardiology after a four chamber view of the heart in the local hospital indicated an abnormality. They were examined by an Advanced Technical Laboratories Mark 4 sector scanner and a Hewlett Packard 77020A phased array scanner with 5 MHz transducers. Both machines can be used for Doppler evaluation of intracardiac velocities and the Hewlett Packard 77020A can be used for colour flow mapping.

The parents were extensively counselled about the experimental nature of the procedures to be attempted. The therapeutic procedures were performed in the Fetal Medicine Unit at Guy's Hospital with an Acuson 128 ultrasound system for visualisation of the needle course and manipulation of the guide wires and balloon catheters. Eighteen gauge transabdominal chorionic villus sampling needles were used (Rockett, London). For needle puncture of the umbilical cord and cardiac chambers we used a freehand ultrasound guided method adapted from the technique first described by Daffos *et al.*⁵ The maternal skin was infiltrated by local anaesthetic before each needle insertion. Intra-uterine and intracardiac pressures were measured by a sterile system of fluid-filled tubing connected from the hub of the needle via a solid state transducer to a Siemens Mingograf by a previously described method.⁶ In the first procedure we used a USCI coronary balloon catheter, diameter 2.5 mm when inflated. In the second case a 3.5 mm diameter balloon was custom made by NuMed.

CASE 1

This 23 year old woman (para 2) was initially referred at 22 weeks' gestation. The left ventricle was of normal size for the gestation but was contracting poorly. There was little discernible forward flow into the left ventricle or aorta. The left ventricular wall showed

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increased echogenicity. Colour flow mapping showed a patent foramen ovale. The parents were advised of the likely poor prognosis but the mother elected to continue the pregnancy. A repeat scan at 28 weeks' gestation showed that the left ventricle had not changed in size and was therefore small for the gestation by this time. There was no flow detectable in the ascending aorta and the atrial septum was now closed. The intrauterine procedure took place a few days later. The mother was sedated with intravenous pethidine. The fetal position was initially unfavourable with the spine anterior, but spontaneous fetal movement resulted in a more suitable position with the cardiac apex positioned laterally about an hour later. On the first needle insertion, the right ventricle was entered and a pressure tracing recorded. The needle was then withdrawn and the left ventricle entered through the left ventricular free wall. The balloon catheter was passed down the needle but the guide wire could not be made to cross the aortic valve. At this time at one point, the needle crossed the aortic valve. We thought that the route of approach was not sufficiently direct so a new needle insertion was attempted. This entered the heart at the apex and in line with the origin of the aorta. However, the guide wire was repeatedly deflected off the aortic valve to pass retrogradely through the mitral valve. Three episodes of fetal bradycardia occurred during the procedure; these were reversed with intracardiac isoprenaline. The attempt was considered to have failed and therefore was abandoned at this stage. The fetus was active and the heart was in sinus rhythm at the end of the procedure but death occurred during the next 24 hours.

Postmortem examination showed a single puncture mark on the anterior chest wall. There was 7 ml of blood in the left pleural cavity and 3 ml in the right. There was no blood in the pericardial cavity. The heart showed a moderate degree of left ventricular hypoplasia, a small stenotic mitral valve, and endocardial fibroelastosis of the left ventricle. The ascending aorta was small for gestational age. The valve was formed by a thin but complete membrane that had been raggedly torn, apparently during the interventional procedure and not at necropsy. The foramen ovale was not patent.

CASE 2

A 41 year old woman (para 2) was initially referred at 30 weeks' gestation. A normal fetal karyotype had been found on chorionic villus sampling at nine weeks' gestation. At a routine scan in the local hospital at 18 weeks' gestation, the four chamber view was passed as normal. The second scan at 30 weeks was to check placental position but the fetal heart seemed to be abnormal and the patient was referred to Guy's Hospital. The left ventricle was poorly contracting and thickwalled with increased echogenicity of the lining. There was, however, some forward flow through the mitral valve and the atrial septum was patent. The aortic valve seemed thickened and restricted in motion with

the velocity of flow increased to 150 cm/s.

The first interventional procedure took place a week later at 31 weeks' gestation. The fetal orientation was favourable from the start such that a direct approach from the apex could be made on the first attempt. A guide wire was advanced through the needle and across the aortic valve under direct view. The balloon catheter was then inserted and inflated. The balloon, however, seemed to be in the left ventricular outflow tract and not in the valve orifice itself when inflation took place. Retrospective review of the videotape recorded at this time confirmed this. The balloon burst in situ, and when it was being withdrawn through the needle it impacted on the edge of the needle and was torn from the main body of the catheter. The needle and the portion of catheter within it were then withdrawn as one. The fetal heart remained in sinus rhythm throughout. No change in flow could be detected across the aortic valve after the procedure and the attempt was deemed unsuccessful. Subsequently the balloon and wire could be seen on ultrasound examination to be impacted between the left atrium and the septal left ventricular wall below the outflow tract. It was not causing any obstruction.

A second attempt was made 10 days later at 33 weeks' gestation. The left ventricular apex was entered on the second needle placement. A guide wire was entered through the needle and passed easily across the aortic valve. After we established that the valve could be crossed with the guide wire the wire was withdrawn and the custom made balloon catheter was then inserted. It was inflated twice within the valve orifice. The catheter and needle were then removed simultaneously. The fetal heart rate was normal throughout except for a transient bradycardia at balloon inflation that did not require treatment. Fetal movement did not interfere with the procedure. The technical success of the procedure appeared to be confirmed by an improvement in aortic flow shown by Doppler echocardiography; the velocity of flow fell from 150 cm/s to 120 cm/s.

One week later, the mother went into spontaneous labour and was delivered in the local hospital. The neonate was male, had Apgar scores of 7 and 9 at 1 and 5 minutes and weighed 2.98 kg. He was transferred to Guy's Hospital on facial oxygen and a prostaglandin infusion. The Doppler gradient across the aortic valve on admission was 36 mm Hg. The reduced function and echogenicity of the left ventricle were confirmed. There was a large ductus arteriosus. The remnants of the balloon used for the first procedure were seen in the left ventricular outflow tract close to the septal wall. Balloon dilatation with a Schneider-Shiley catheter passed retrogradely from the femoral artery was successful when he was 18 hours old. An aortogram showed mild distal aortic arch hypoplasia but seemed to exclude aortic coarctation. There was no increase in Doppler estimated velocity around the aortic arch.

The infant required continuous ventilation and circulatory support. Left ventricular function initially seemed to improve but by day 10

it was dyskinetic. Renal failure developed and was treated by peritoneal dialysis. By day 27, however, there was no recovery of renal function and the neonate showed impaired hepatic and left ventricular function. On Doppler examination there was no gradient across the aortic valve or around the aortic arch at this time and the duct had closed. After conferring with the parents, intensive care was stopped on day 28 and the neonate died.

A postmortem examination showed that the left ventricular wall was thickened and lined with fibroelastosis. The remnants of the original balloon catheter and guide wire lay in the left ventricular outflow tract but were firmly anchored in the myocardium. They were not causing an obstruction. The aortic valve was dysplastic though the cusps were cleanly split along the commissures. The diameter of the aortic root at the origin was 8 mm, with a maximum diameter of 11 mm in the ascending aorta. The aortic arch narrowed to 5 mm with the suggestion of a small, distal, discrete coarctation shelf when the arch was opened.

Discussion

Aortic valve disease can be diagnosed with accuracy by echocardiography before birth. The diagnostic features include a poorly contracting left ventricle, aortic valve thickening and restriction, a varying degree of left ventricular hypertrophy and echogenicity, and abnormal Doppler flow characteristics in the left heart. There may be little or no detectable flow into or out of the left side of the heart. The atrial septum is often closed at the initial examination, or closes as gestation advances, as shown in one of our patients.

In our prenatal series of 12 continuing pregnancies some cases did not survive until delivery and a further group did not live to reach either the catheter laboratory or surgery. Those who survived to undergo relief of aortic obstruction all subsequently died of endocardial fibroelastosis or left ventricular hypoplasia. In the face of this mortality we considered prenatal treatment.

The technique of transuterine cardiac puncture for fetal blood sampling or fetal transfusion is well established in specialised obstetric practice. During the procedures all the drugs normally necessary for resuscitation were available in doses appropriate to the fetal size. The balloon catheters used were a modification of those used in postnatal life and the procedure for positioning the guide wire and balloon within the valve was similar to that practised in children. Thus in the prenatal procedure we described we combined two established techniques and brought together the expertise of the two specialties.

It is important to consider the possible hazards to the mother and the fetus and to weigh them against the potential benefit of the procedure. Sterile techniques were used for insertion of needles and both mothers were given prophylactic antibiotics before and for 24 hours after the procedure. Maternal discomfort can be minimised by premedication and local

anaesthetic at the site of needle puncture. It is possible that premature labour may be precipitated and this would require active treatment. There seems to be little associated tissue damage to the fetus. Haemorrhage from the puncture site may have caused the intrapleural collections found at necropsy in the first case though there was no intrapericardial blood. The fetal position of case 1 necessitated crossing the lung fields with the needle whereas the position of case 2 allowed a direct apical puncture and no intrapleural fluid was seen on ultrasound in the days after the procedure. In case 1, the needle itself apparently tore the aortic valve but this could be an intended first step when the guide wire fails to cross the aortic valve. In the second case, there was no evidence of a needle puncture site in the newborn at delivery one week after the interventional procedure nor was intracardiac trauma found at necropsy. The detachment of part of the balloon and its impaction in the ventricular wall in the second case was a technical error but one which may be avoided in the future by improvements in the design of the balloon catheters and needles.

It may be that patients with aortic stenosis who are suitable for this procedure require to be carefully selected. For example, where there is already established severe endocardial fibroelastosis or where the left ventricle is failing to produce an increased velocity of flow across the stenotic valve, ventricular damage may already be irreversible. Once the atrial septum is closed, relief of left ventricular obstruction alone may not be sufficient to maintain left ventricular growth. Where the conditions seem favourable for balloon dilatation of the aortic valve, it may also be necessary to dilate the mitral valve because there is often considerable inflow obstruction associated with aortic valve disease.

In summary, case 1 proved that delivery of the balloon to the left ventricle is possible in utero and case 2 that balloon dilatation of the fetal aortic valve is technically possible. Further experience with the technique will clarify which patients will benefit most from this form of treatment. Though the place of this form of treatment in the varying options for management of this condition has yet to be established, this preliminary step to prenatal treatment seems promising.

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